Hexosaminidase A B

General:

Hexosaminidase is an autolysis enzyme involved in the hydrolysis of several molecules containing hexose. Beta-hexosaminidase A and betahexosaminidase B play a critical role in the brain and spinal cord (central nervous system). These enzymes are found in lysosomes, which are organelles in cells that break down toxic substances and act as recycling centers. Within lysosomes, the enzymes break down fatty compounds called sphingolipids, complex sugars called oligosaccharides, and molecules that are linked to sugars (such as glycoproteins). In particular, betahexosaminidase A forms part of a complex that breaks down a fatty substance called GM2 ganglioside.

Deficiencies cause an inability to properly hydrolyze certain sphingolipids, causing these lipids to accumulate over time in lysosomes. Disorders of hexosaminidase are responsible for the following conditions: Tay-Sachs disease (type A only), lysosomes fill with GM2 gangliosides; Sandhoff disease (types A and B), lysosomes cannot break down globosides.

Indication:	Suspicion of gangliosidosis
Preanalytics:	Dried blood spots on filterpaper, store and dispatch at room temperature, not freezing, thermal effects can reduce enzyme activities.
Material:	Dry spot blood (filter card)
TAT:	2 weeks*
Method:	ENZ
Units:	nmol/spot
Ref.- range:	0.6 -2.4

For complete list of laboratory test offered at Freiburg Medical Laboratory, please visit http://www.fml-dubai.com/parameter-listings/

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